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Primary stenosis of the sphincter of Oddi : literature review and case presentations

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To the memory of my mother, who filled
my past
And to my wife, who is my present

Acknowledgements

I would like to express my sincerest gratitude to Dr. Howard Spiro for his guidance and inspiration. He assisted me in many ways with his pedagogical acumen and special wit. I would also like to thank Dr. Jerry Avella and Dr. John Dobbins for their invaluable assistance in helping me gather the clinical data.

HISTORICAL REVIEW

Stenosing lesions of the sphincter of Oddi have become more frequently described since their initial recognition by Carl Langenbuch in 1884. In a paper by Grage, et al.,[9] Langenbuch is quoted as suggesting transduodenal division of the "diverticulum" of Vater in cases of cicatricial stenosis from chronic inflammation. McBurney in 1882 removed an impacted stone from the ampulla of Vater after doing a sphincterotomy. Then in 1894, Kocher utilized the same technique for removing stones from the distal end of the common bile duct but approximated the cut mucosal edges of the bile duct and the duodenum. He called this a "choledochoduodenostomia interna" or sphincteroplasty.

The idea of causal relationships between distal biliary tract obstruction and acute pancreatitis was established by the work of Opie in 1901. This initiated the "common channel" theory, whereby an impacted stone in the ampulla is deemed the cause of acute pancreatitis. This led Archibald in 1913 to suggest sphincterotomy for the relief of increased biliary pressure.

Much literature has been produced concerning the possible etiologies of obstruction of the distal common bile duct. Berg in 1922 proposed the idea of sphincteric dysfunction leading to the development of biliary stones which would secondarily result in changes in the sphincter. Del Valle in 1928 proposed that spasm of the sphincter

would be aggravated by fibrosis, which in effect was the result of long term spasm. He also believed that it was possible to diagnose a stenotic sphincter, preoperatively, by ascertaining the area of radiation of the pain. His clinical experience showed that pain due to common bile duct obstruction, from either spasm or fibrosis of the sphincter of Oddi, typically radiated to the level of the first lumbar vertebra and also to the left scapular angle. The pain of biliary colic, on the other hand, radiated to the mid-thorax, right hemithorax, and right shoulder tip. Del Valle also observed in 1930 that some patients were without relief of the pain after cholecystectomy and oftentimes had more severe pain. He attributed this to the loss of the decompressive action of the gallbladder.

The idea of spasm of the sphincter of Oddi being the cause of pain did not receive much attention until the published studies of McGowan, Butsch, and Walters in 1936. They demonstrated a causal relationship by measuring the pressure in the common bile duct through a T-tube in a cholecystectomized patient. Administration of morphine sulphate resulted in an intraductal pressure rise as high as 160 mm of water with a concomitant attack of pain similar to that experienced prior to cholecystectomy. Doubilet and Mulholland[7] reported later in 1956 the relationship between rising intraductal pressure and emotional upset and unpleasant conversation. (See Fig. 1.) They concluded that

either intermittent or continuous spasm caused by tension and emotional frustration led to reflux of bile into the pancreatic duct, resulting in progressive destruction of the pancreas.

Studies concerned with the forceful dilatation of the sphincter of Oddi and its effects were done by Branch, Bailey, and Zollinger in 1939. They used dogs in their study and found moderate to marked residual scarring in one half of the dogs after the trauma of dilatation. They also observed hepatic duct dilatation in the affected dogs. Another one of their findings was that dilatation did not maintain the patency of the lumen of the papilla. Experiences of Allen and Wallace, also in 1939, seemed to dispute the experimental results if careful dilatation was less than the diameter of the common bile duct. The use of Bakes dilators became routine in common duct explorations after its introduction by Chute and Allen. Doubilet and Mulholland felt, however, that passage of a stone or a large probe would result in fibrosis.

Fibrosis, as an associated histological finding, has been extensively reported in the literature pertaining to cases of stenosis of the sphincter of Oddi. Mahorner in 1945 reported three cases of fibrosis and, later in 1949, Trommald and Seabrook[25] were to report eight cases of benign fibrosis of the sphincter. In 1953 Cattell and Colcock drew attention to the importance of fibrosis as an

etiological factor in the "postcholecystectomy syndrome." They noted that, in a series of 100 patients with fibrosis, 81% of their patients had undergone a previous cholecystectomy without relief of symptoms[5].

The persistence of the symptoms after cholecystectomy in the postcholecystectomy syndrome has been theorized to have numerous etiologies. Cystic duct stump, neuroma, disturbed innervation of the biliary duct, recurrent pancreatitis, reflux of bile and pancreatic fluid, and spasm and stenosis of the sphincter of Oddi, have all had their proponents.

Riddell and Kirtley,[20] in their series of 31 patients, reported 16 patients with the postcholecystectomy syndrome, seven of whom had no calculi reported at the primary operation. An additional eight patients were reported to have primary or idiopathic stenosis of the sphincter of Oddi. This group was found at operation to not have evidence of liver, gallbladder, common bile duct or pancreatic disease. They also had the additional distinction of not having undergone previous biliary surgery.

What may be the first reported case of primary stenosis of the sphincter of Oddi is that published by Summers[24] in 1900. The case was that of an 18 year-old girl with a five-year history of intermittent attacks of "hepatic colic." The attacks became more severe during the first four years and were often accompanied by jaundice. The patient appeared

very jaundiced on physical examination and had considerable epigastric and right upper quadrant pain. The patient was also noted to have hepatosplenomegaly. On laparotomy, it was noted that the gallbladder and the common bile duct were markedly distended. Stones could not be palpated in either the cystic or the common bile duct. Summers noted a stricture at the distal end of the common bile duct when he attempted to pass a probe into the duodenum. Choosing not to dilate the stricture, he then proceeded to anastomose the opened gallbladder to the wound of the duodenum. Postoperatively, the patient did well except for an infection which subsided, and subsequently reported a weight gain and no recurrence of pain.

Retrospectively, one cannot be at all certain that this case represents primary stenosis, in that the anatomical lesion is vaguely described as an "evident stricture near the duodenal end of the duct." This case does illustrate, however, the oftentimes perplexing nature of biliary type pain in the absence of calculi.

The clinical entity of primary stenosis of the sphincter of Oddi had not been widely recognized until the paper by Riddell and Kirtley. Nardi in 1966 considered the entity of idiopathic pancreatitis in evaluating patients with pancreatitis of unknown etiology. The prevalence of primary stenosis is very low, as evidenced by the total number of presumptive primaries in Table I, drawn from the literature

under review. The proviso, presumptive, is used because most of these authors do not fully define idiopathic or primary stenosis.

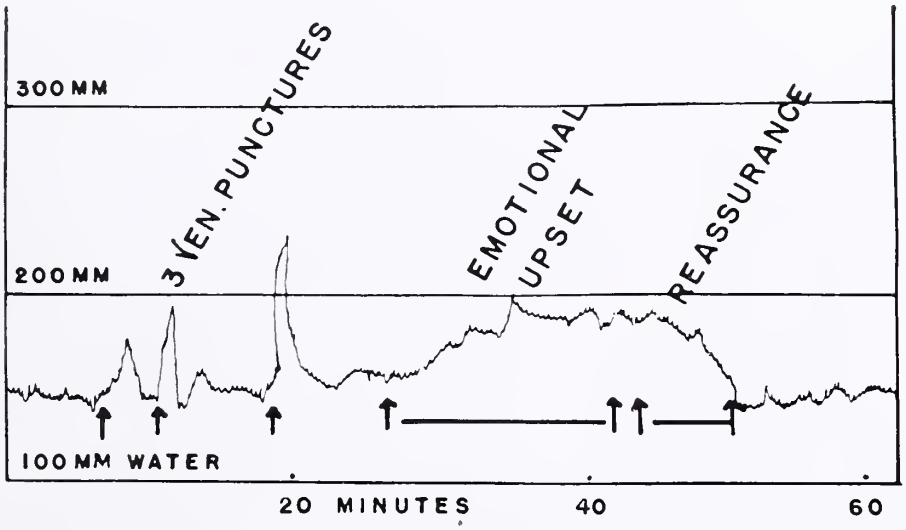


Fig. 296. Kymographic recording of the tonus of the sphincter of Oddi in the human. Note transitory spasm produced by the pain of venipunctures and the persistent spasm accompanying an emotional upset. Reassurance resulted in the disappearance of the spasm.

Fig. 1.

TABLE I

Author	Date	Tot. in Series	# Primaries	% Primaries
Trommald[25]	1950	8	2	25
Riddell[20]	1959	31	15	48
Hendren[11]	1965	15	3	20
Nardi[15]	1966	67	17	25
Acosta[1]	1967	38	10	26
Nardi[16]	1970	1	1	
Puente[19]	1970	66	1	1.5
Stefanini[21]	1974	712	36	5
Haff[10]	1975	23	4	17
	TOTAL	961	89	9

Anatomy of the Sphincter of Oddi

The normal anatomical architecture of the sphincter of Oddi involves the following:[1,4,14,17]

Papillary Mucosa - The mucosa may have long folds and forms an irregular series of wavelike structures. According to McPhedran, et al.[14], these finger-like projections exist either as single stalks or as transverse folds that wave freely in the lumen of the ampulla and pancreatic duct. V. Patzelt, as quoted by McPhedran, suggests that these fronds prevent reflux from the duodenum into the common bile and pancreatic ducts. Histologically, the epithelium is tall and cylindrical with numerous goblet cells.

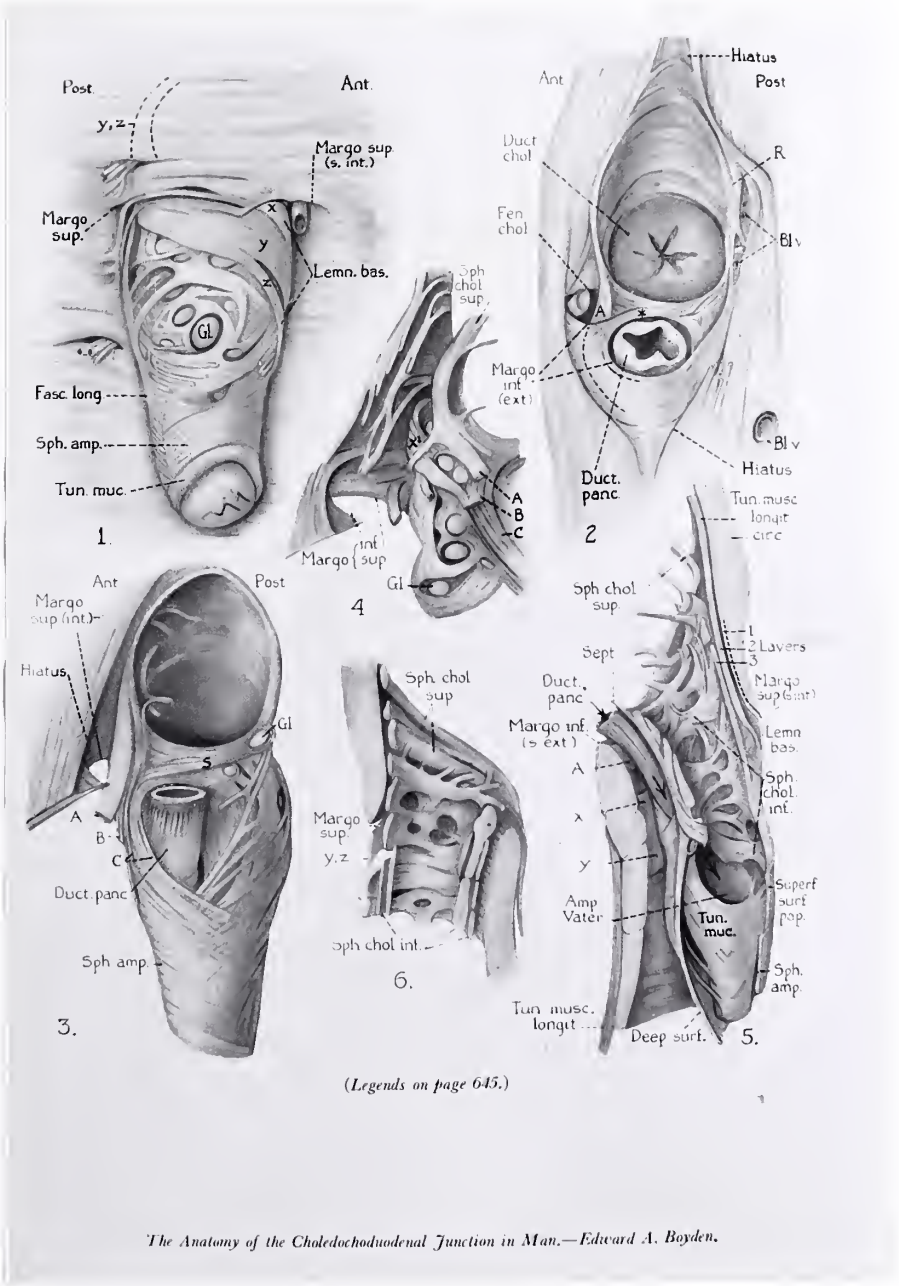
Papillary Submucosa - It contains numerous mucosal glands, especially at the depths of the mucosal folds. The muscularis mucosae is poorly formed, consisting of only a few isolated muscle fibers which may ascend into the mucosal folds. Loose connective tissue surrounds the glands and, when polarized light is used, thick, isolated collagen fibers are seen.

Sphincter of Oddi - The musculature lies between the ampullary submucosa and the duodenal submucosa. The fibers vary considerably in orientation with respect to the lumen because of the presence of entrapped deep submucosal glands. (See Fig. 2.) The smooth muscle layer may be more orderly near the junction of the common bile

duct and the duct of Wirsung, where the muscle lays parallel to the lumen. One may observe randomly distributed bundles of fibers, frequently oriented around the entrapped glandular elements near the papillary orifice. There is sparse connective tissue which appears on polarized light microscopy as a few thin, isolated, doubly refractile lines. The walls of the normal common bile duct are composed of dense connective tissue which may be interpreted as being pathologic.

Duodenal Submucosa - It is part of the ampullary wall and contains only a few Brunner's glands surrounded by loose connective tissue.

Duodenal Mucosa - The epithelium is continuous with the epithelium of the ampulla. A well developed muscularis mucosae is present up to their junction.



(*Legends on page 645.*)

The Anatomy of the Choledochoduodenal Junction in Man.—Edward A. Boyden.

Fig. 2. Reprint of Boyden's drawings of the muscles of the sphincter of Oddi.

Materials and Methods

The records of 52 patients who had undergone sphincteroplasty within the past 22 years on the university surgical service were reviewed. Seven patients (13.5%) are considered to represent primary stenosis of the sphincter of Oddi. The criteria used to establish this clinical entity are: (1) the inability to find calculi at the primary and, if applicable, the secondary operation; (2) inability to demonstrate calculi preoperatively and postoperatively on x-ray; (3) a negative history of alcohol abuse; (4) the lack of previous surgical manipulation of the biliary tree, such as T-tube placement during the primary operation; and (5) inability to pass a #3 Bakes dilator through the sphincter of Oddi. The case histories of the patients follow.

Case #1, V.S. (unit #92-45-14)

The patient is a 15 year-old Puerto Rican male, who presented to his family physician with hyperthyroidism. The patient was treated with Propylthiouracil and soon after became jaundiced, accompanied by light-colored stools, dark urine, and nosebleeds. The patient then developed pruritus and ankle edema, therefore the diagnosis of serum hepatitis was entertained. On physical examination, the patient was noted to be deeply icteric and goitrous, and displayed hepatomegaly. The patient's primary problem was diarrhea on admission to Yale - New Haven Hospital. Evaluation included a small bowel biopsy and an upper GI, which were essentially normal. Laboratory studies were significant for thyroid indices consistent with hyperthyroidism; liver function studies showed a total bilirubin of 35.5, direct bilirubin of 17.5, SGOT of 340, and an alkaline phosphatase of 144. The patient underwent ERCP which revealed obstruction at both the pancreatic and common bile ducts. (See Fig. 3.) Also noted was a stricture in the proximal portion of the biliary tree. The patient then became febrile and stuperous, indicating deterioration of his condition.

The patient then underwent exploratory laparotomy in March 1975, when the presence of multiple, soft stones and thick bile in the gallbladder was noted. The sphincter of Oddi was able to pass a #4 Bakes dilator

but unable to allow a #5. A sphincteroplasty was done in addition to a cholecystectomy, liver biopsy, lymph node biopsy, and pancreatogram, which was normal.

One month postoperatively, the patient developed diarrhea and a fever to 101. It was felt that the patient had developed pancreatic insufficiency. The patient was then started on Viokase with subsequent improvement. The patient also underwent an upper GI which demonstrated patency of the sphincteroplasty but no reflux of barium. The patient was gaining weight and doing well post-operatively.

Comment:

This case is atypical in its presentation, in that the patient presented with jaundice and light-colored stools but without right upper quadrant or epigastric pain. The argument could be made that this case does not represent primary stenosis of the sphincter of Oddi. However, the surgeon noted the existence of a congenital stricture even though it was able to accept the passage of a #4 Bakes dilator. The findings at ERCP indicate distal obstruction with the greatly dilated pancreatic and common bile duct. The presence of multiple, small, soft stones was probably due to bile stasis, as evidenced by the liver biopsy which showed "marked cholestasis..." Biopsy of the sphincter revealed fibrosis. (See Fig. 4)

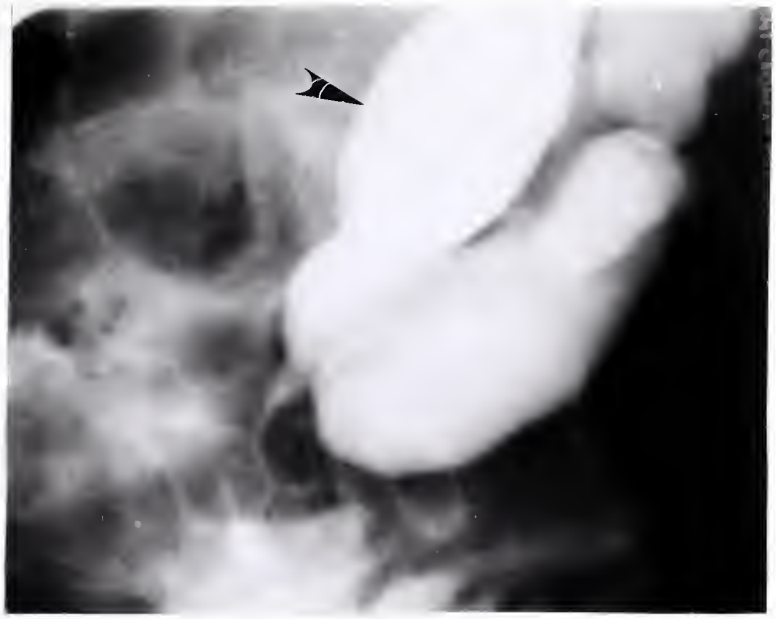


Fig. 3. ERCP from case #1. Note the grossly dilated pancreatic and common bile ducts. (Arrows)

Fig. 4. Biopsy of ampulla of Vater from case #1. Duodenal mucosa (small arrows) and sphincteric musculature (large arrows).

Case #2, R.L. (unit #95-11-42)

The patient is a 41 year-old white male with a history of recurrent acute pancreatitis. The patient's first attack was in 1959, which was characterized by midepigastric pain, nausea, vomiting, and bloating. The patient denied radiation of the pain. The patient was hospitalized at the Ochsner Clinic in New Orleans, where laboratory documentation of pancreatitis was made. The patient underwent sphincterotomy in 1960 and 1963. It was noted by the surgeon that no stones were found and studies made of the biliary tree were normal at both operations. The only abnormality noted was stenosis of the sphincter of Oddi. The patient denied a history of biliary tract disease, alcohol abuse, hemolysis, or a family history of similar problems.

The patient enjoyed a hiatus of good health until December 1975, when he again developed abdominal pain, nausea, vomiting, and elevated serum amylase. Intravenous cholangiogram showed the possibility of calculi in either the common bile duct or the pancreatic duct. An upper GI revealed no reflux of barium. He then underwent an ERCP which showed "a normal common bile duct, mild narrowing in the neck of the main pancreatic duct, and two probable stones in the pancreatic duct." (See Figs. 5 & 6.) Following this procedure, the patient developed acute pancreatitis with an amylase of 4180 and a lipase of 57.6,

which was treated with antibiotics and nasogastric suction. He remained febrile but without peritoneal signs. There was no evidence of a pancreatic abscess or a pseudocyst noted on diagnostic ultrasound. The patient then underwent an exploratory laparotomy in March 1976. Intraoperative pancreatogram revealed a dilated pancreatic duct. A sphincteroplasty was done on the common bile and the pancreatic ducts. The patient has done reasonably well except for an undocumented episode of epigastric pain while on vacation.

Comment:

This patient's pancreatitis was probably due to stenosis of the sphincter of Oddi. Restenosis of the sphincterotomy is highly probable in light of the patient's history of restenosis. The formation of the calculi, however, is open to speculation.

Fig. 5. ERCP from case #2 showing two stones in the pancreatic duct (arrows).

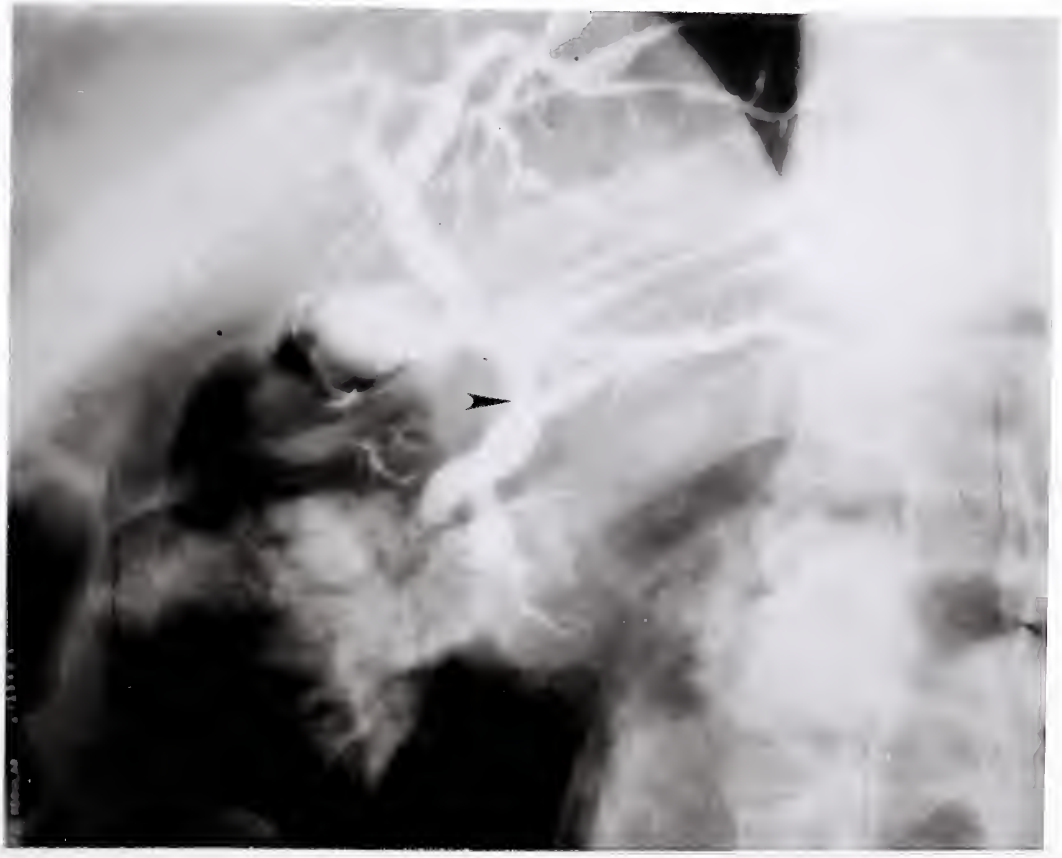


Fig. 6. ERCP from case #2 showing a normal common bile duct (arrow).

Case #3, C.W. (unit #69-20-09)

The patient is a 32 year-old white female who worked as a psychiatric nurse and had an eight-year history of recurrent right upper quadrant and epigastric pain. The patient admitted a possible correlation between emotional stress and the pain, which she described as being colicky in nature with radiation to the back and occasionally to the chest. The temporal aspect of the pain was related to the late afternoons or evenings, and it usually awakened her at night. The patient denied any causal relationship to food, position, or activity, but did obtain relief from antacids. She also denied fever, chills, weight loss, nausea, vomiting, or jaundice. The patient underwent an upper GI, oral cholecystogram, endoscopy, and ERCP, which were all normal. (See Figs. 7 & 8.) Chemical pancreatitis complicated ERCP with an increase in serum lipase from 1.7 to 11.9 and serum amylase from 197 to 916.

At laparotomy, it was noted that there were no calculi in the gallbladder or in the common bile duct. It was also noted that the sphincter of Oddi would not accept the passage of a #3 Bakes dilator, at which point a sphincteroplasty was done. Postoperatively, the patient does not seem to be improved by this procedure, in that she reports recurrence of right upper quadrant pain.

Comment:

Although recurrence of this patient's symptoms may suggest the lack of benefit, the fact remains that there was stenosis of the sphincter of Oddi without any other associated pathology. The existence of some psychogenic overlay may be clouding the issue of this patient's pain.

Fig. 7. ERCP from case #3 showing a normal common bile duct and a normal pancreatic duct (arrows).

Fig. 8. ERCP from case #3 showing more proximal portion of the pancreatic duct.

Case #4, L.E. (unit #57-73-63)

This patient's symptomatology began in 1953 while in prep school, where he fasted to bring his weight down for a wrestling match. He then ate a large meal which initiated an episode of abdominal pain. Upon admission to the school infirmary, his pain subsided. Several years later, while a freshman in college, the patient again fasted for a wrestling match with a large meal following, resulting in severe right upper quadrant pain which radiated across the midepigastrium and around to the left costovertebral angle. He was hospitalized with documented acute pancreatitis at the Massachusetts General Hospital, where he underwent an intravenous cholangiogram which showed a normal gallbladder and a normal intrahepatic duct system. The distal portion of the common bile duct was not visualized. The patient also underwent a carefully monitored secretin test which was normal. He continued to have recurrent attacks of acute pancreatitis until July 1959, at age 22, when he underwent exploratory laparotomy at the University of Virginia Medical Center, where he was a medical student. The patient was found to have had a markedly stenotic and fibrotic sphincter which would not admit the passage of a #3 Bakes dilator. The common bile duct was also noted to be narrowed. No calculi were seen and a sphincterotomy was performed with the gallbladder left in.

The patient did relatively well with only minor discomfort and no attacks of pancreatitis until 1963. Serum amylase and lipase levels peaked at 1375 and 9.4 units respectively. Persistence of the symptoms of right upper quadrant pain with radiation to the right subscapular area led to reexploration in 1966. He underwent a cholecystectomy and a sphincteroplasty because of restenosis. The patient was noted to have extensive calcification seen on x-ray but continues to do well as reported by Spiro.

Comment:

This patient's initial presentation was reported in 1956 by McDermott, et al.[13] It was postulated that the "prolonged period of starvation and dehydration might have resulted in a thick, inspissated secretion in the ductal system of the pancreas and that the intense stimulation of the pancreas" from a large intake might well have produced an increase in pressure in the pancreatic ducts, with the rupture of multiple acini, causing the pain of acute pancreatitis. The relief brought on by sphincterotomy and subsequent restenosis with pain clearly points to the sphincter of Oddi as the cause of his pain.

Case #5, J.J. (unit #65-42-31)

This 25 year-old graduate student was first seen at Yale - New Haven Hospital in November 1965, for abdominal pain and vomiting. The patient had been initially diagnosed by his local physician as having acute pancreatis in July 1964, when it was noted that he presented with dull, epigastric pain. The pain did not radiate and was aggravated by food. The patient denied a history of alcohol abuse. An oral cholecystogram showed a poorly functioning and visualizing gallbladder; an upper GI was negative. He had a history of hospitalizations as a young child for abdominal pain and constipation of unknown etiology.

Subsequent studies at Yale revealed a normal upper GI, an intravenous cholangiogram showed no calculi, but calcification was noted adjacent to the termination of the common bile duct. During the course of hospitalizations for attacks of acute pancreatitis, the patient's serum amylase and lipase peaked at 600 and 5.3, respectively. The patient underwent exploratory laparotomy in October 1968, because of persistence of his symptoms. No calculi were seen, but it was noted that the common bile duct was small and the sphincter of Oddi stenotic. A cholecystectomy and a sphincteroplasty were done. The patient was re-explored one year later for a slowly expanding abdominal mass which was found to be a pseudocyst. The patient has done well as reported to me by Dr. Spiro.[22]

Comment:

This case and case #4 were both summarized by Strum and Spiro in 1971.[23] Spiro's subsequent reports from this patient's physician reveal that he is doing very well and is eating a normal diet. Except for the pseudocyst which complicated the patient's postoperative course, the relief of his pain would implicate a stenotic sphincter of Oddi.

Case #6, D.A. (unit #97-53-34)

The patient is a 36 year-old white female with a ten-year history of right upper quadrant occurring in two to three-year intervals. The patient was initially hospitalized ten years prior for epigastric pain which was treated with a bland diet and antacids. She noted the increased frequency and severity of her abdominal pain for about six months prior to her admission to Yale - New Haven Hospital. It was characterized as occurring every two to three days and being most severe at night, arousing her from a sound sleep. The paroxysms of pain would last for about six hours and were unrelieved by food, Maalox, or activity, but she found relief in ingesting Percodan. The patient underwent an upper GI, barium enema, sigmoidoscopy, oral cholecystogram, and panendoscopy, which were all negative. The patient's family history was remarkable for the occurrence of gallstones in her father and sister.

At presentation to Yale - New Haven Hospital, the patient was noted to have a normal physical examination. She then underwent ERCP, which was essentially normal and failed to demonstrate delayed drainage. (See Figs. 9 & 10.) It was felt that the patient had stenosis of the sphincter of Oddi and would benefit from a sphincteroplasty. At laparotomy, the patient was noted to have an acalculous gallbladder and a stenotic sphincter of Oddi, which would

not accept the passage of a #3 Bakes dilator. A sphincteroplasty and cholecystectomy were done and the patient did well postoperatively. Pathologic diagnosis was that of chronic acalculous cholecystitis and fibrosis of the ampulla of Vater. (See Fig. 11.)

Comment:

The possibility of stenosis of the sphincter of Oddi occurring in her father and sister has not been explored. The occurrence of stenosis leading to bile stasis with the formation of stones is conceivable but difficult to prove. The inability to demonstrate calculi with an oral cholecystogram or ERCP makes the passage of a stone highly unlikely. The length of followup of this patient is not long enough to conclude whether she benefited from this procedure.

Fig. 9. ERCP from case #6. Arrows are pointing to the gallbladder and the common bile duct.

Fig. 10. ERCP from case #6 showing the proximal portion of the pancreatic duct.

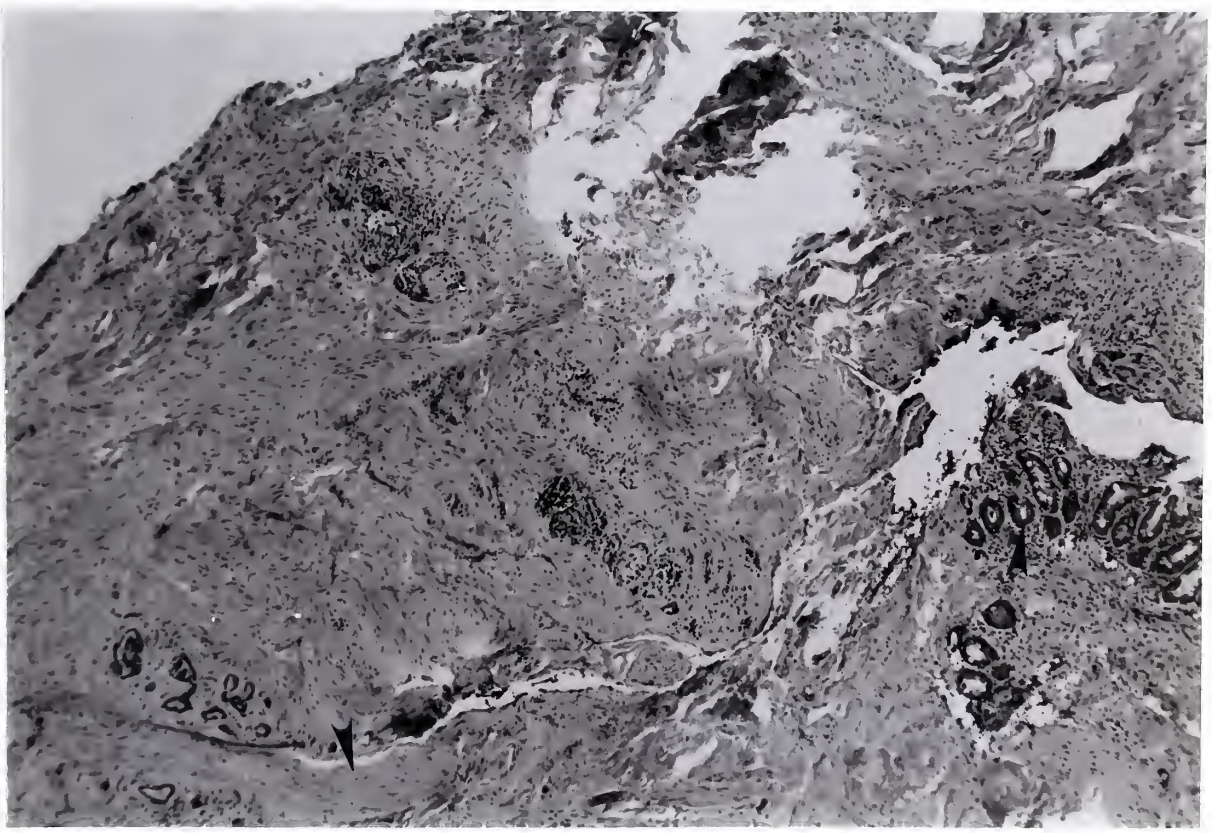


Fig. 11. Biopsy of the ampulla of Vater from case #6. Arrows indicate muscle fibers and Brunner's glands.

Case #7, V.D. (unit #86-80-38)

The patient is a 47 year-old white female, who was referred to Yale - New Haven Hospital because of recurring attacks of pancreatitis. She is reported to have had a cholecystectomy thirteen years prior because of cholesterolosis without calculi. The patient did well until 1972, when she noted the onset of pain in the upper abdominal region which radiated to her back. The patient had a negative evaluation at another institution. In the interval of time between her evaluation at the other institution and her presentation at Yale, she noted the continuance of epigastric pain with radiation to the back, occurring about two to three times per month. She also noted associated fevers up to 103, but denied the appearance of jaundice, changes in stool or urine color, or vomiting. A repeat upper GI and barium enema were normal. The patient denied alcohol abuse or food intolerance.

Presentation at Yale revealed a normal physical examination. Laboratory studies showed a normal serum amylase and lipase, an SGOT of 31, LDH of 370, and an alkaline phosphatase of 26. An attempt to do an ERCP was unsuccessful. It was then elected to do a laparotomy because of the severity of the symptoms. At laparotomy in June 1973, it was noted that the ampulla would not accept the passage of a #10 red rubber cather but did accept the passage of a #3 Bakes dilator, afterwards. A transduodenal

sphincteroplasty was done and the patient did well post-operatively. One year postoperatively, it was noted that the patient was without the original pain.

Comment:

The finding of cholesterolosis at the primary operation may be presumptive evidence for bile stasis secondary to distal obstruction. Preston has suggested that the finding of cholesterolosis cholecystitis "should alert the surgeon to the possibility of stenosis or dyssynergia of the sphincter of Oddi."[18] The exact size of the ampulla is not accurately reported, but the impression was stenosis. Riddell and Kirtley report having two patients in their series with cholesterolosis of the ampulla. One patient is described as being in the category of having cholelithiasis but no mention is made of the other patient. The patient presented would probably be classified as having the postcholecystectomy syndrome.

Results:

Analysis of the patients presented show that four patients had signs and symptoms of acute cholecystitis and three patients had recurrent pancreatitis. (See Table II.) The male:female ratio was 4:3, greater than what one would expect, but this series may be too small to draw definite conclusions. All of the patients in this series had undergone either an oral cholecystogram or an intravenous cholangiogram with negative results, except in case #2, where the possibility of calculi in either the common bile duct or the pancreatic duct was raised. The presence of calculi in the pancreatic duct was confirmed by the use of Endoscopic Retrograde Cholangiopancreatography (ERCP). ERCP was used in four patients, diagnosing obstruction in only one patient, case #1. It was attempted in one patient, case #7, but was unsuccessful.

Patients who presented with the pain of acute cholecystitis were characterized as having right upper quadrant and epigastric pain which sometimes radiated to the back. Only one patient, case #1, was grossly jaundiced and was atypical in his presentation. The patients with pancreatitis were characterized as having right upper quadrant or epigastric pain, with or without radiation. Serum amylase and lipase were elevated and documented in their histories.

All of the patients in this series underwent

sphincteroplasty for attempted relief of their symptoms. Two patients, cases #s 2 and 4, had prior sphincterotomy. It was noted at laparotomy that restenosis had occurred in these patients. Biopsy of the papilla showed fibrosis in all patients in this study.

Six patients, 86%, presented with their symptoms within the second and third decades of life. The other patient, case #7, presented in the fourth decade of life. The average age of onset of symptoms was 24.7 years and the average age at which surgical correction was done was 31.1 years.

Followup on the patients in this series indicates that five patients are doing well at greater than one year postoperatively. One patient has some persistence of pain and another has pain of possibly psychogenic origin. There were no deaths in this series.

TABLE II

Patient	Age (onset)	Presenting Signs	Duration	Lab.Values	ERCP(?)	IVC	OCG	Followup
V.S. (WM)	15 (same)	Jaundice, pruritis, fever	3mos.	T.bili.-35.5 D.bili.-17.5 SGOT -340 Alk. Phos.144 Amylase -nl	Dilated CBD & Panc.duct			No problems @ 1 yr.
R.L. (WM)	41 (29)	Epigastric pain, no radiation, N&V, bloating, fever	6 mos.	Amylase -553 Lipase -6.9	nl. CBD, 2 stones in panc. duct	+	+	No pain @ 1 yr.
C.W. (WF)	32 (29)	RUQ + epigastric pain, radiation to back & chest	3 yrs.	Amylase-197 Lipase-1.7	Normal		+	? psycho-genic pain
L.E. (WM)	22 (18)	RUQ + epigastric pain, radiation to left CVA, fever	3 yrs.	Amylase-1375	Not done	+		Doing well as of 1977
J.J. (WM)	25 (22)	Dull epigastric pain, no radiation	3 yrs.	Amylase-600 Lipase-5.3	Not done	+	+	Doing well as of 1977
D.A. (WF)	36 (26)	RUQ pain	10 yrs.	Amylase-nl Lipase-nl	Normal		+	Too soon for followup
V.D. (WF)	41 (34)	Epigastric pain, radiation to back, fever	1 yr.	Amylase-nl Lipase-1.1	Attempted, not successful			No pain @ 1 yr.

Discussion

In the assessment of idiopathic or primary stenosis of the sphincter of Oddi, one must rule out the other possible causes of the symptom complex. The presentation may be that of acute pancreatitis or of biliary colic, since obstruction of the distal common bile duct may cause problems in either organ system. The presence of calculi at previous biliary surgery, operative procedures on the common bile duct such as T-tube placement, dilatation of the sphincter, and alcoholism all must be considered in assessing distal biliary obstruction. One must also keep in mind the possibility of a neoplastic process as well as metabolic causes of pancreatitis.

In the literature concerning stenosis of the sphincter of Oddi, very little attention has been paid to primary or idiopathic stenosis. The work done by Riddell and Kirtley in looking at cases of primary stenosis, where other possible etiologies have been excluded, led them to the conclusion that stenosis of the sphincter of Oddi was a possible cause of the "postcholecystectomy syndrome." In their group of 16 patients with the postcholecystectomy syndrome, seven patients had no calculi at the primary operation. The criterion used in the present study would have allowed classification of these patients as having primary stenosis, provided there were no operative procedures done on the common bile duct, as outlined earlier.

Riddell and Kirtley also excluded patients with pancreatitis from their study, thereby not attempting to establish any link between idiopathic stenosis and pancreatitis, although it is interesting that two of their patients with idiopathic stenosis did have elevated serum amylase levels.

In 1966, Nardi, et al., evaluated 67 patients with recurrent pancreatitis of unknown etiology. On the basis of history, physical examination, and operative findings, he was able to divide the patients into two groups. The first group was composed of 17 patients with idiopathic pancreatitis, having no evidence of biliary tract disease, either by history, physical examination, or laboratory studies. Within this group, two patients had a papillary biopsy showing fibrosis and two other patients showed chronic inflammatory changes. A grossly normal common bile duct was found in each case. The second group was that of 50 patients with pancreatitis associated with biliary tract disease. The diagnosis was made on the basis of a history of jaundice, increased serum bilirubin, or the finding of a thickened and dilated common bile duct.

Based on this series, Nardi concluded the following: 1)use of the pancreatic evocative test (morphine-prostigmine injection with reproduction of pain or elevated pancreatic enzymes) was useful in the diagnosis of idiopathic pancreatitis and selection for operative

correction; 2) patients with associated biliary tract disease all demonstrated inflammatory changes in the papilla, whereas only 4 of the 17 patients with idiopathic pancreatitis showed pathologic changes; 3) all but two patients with biliary tract disease benefited from sphincteroplasty and, in the idiopathic pancreatitis group, only the patients with a patent, dilated pancreatic duct on pancreatogram benefited from this procedure. This totaled three patients.

The series from Yale discussed in this study includes patients who present with the pain of acute cholecystitis and acute pancreatitis. Major differences exist between the series presented and those of Riddell and Nardi. The evocative test advocated by Nardi was not used as a diagnostic tool. ERCP, however, was used in five patients with varying degrees of success, the rationale being the ability to demonstrate delayed drainage of radiographic contrast material through the sphincter. It proved to be a useful procedure in two patients but was not without its complications in two others, i.e., precipitating chemical pancreatitis, possibly by triggering spasm of the sphincter.

The patients in Riddell's study underwent sphincterotomy for correction of stenosis. The study presented, like Nardi's, utilized sphincteroplasty for correcting stenosis. Riddell, like other authors, claimed good success with sphincterotomy. The series presented, on the

other hand, includes two patients in which sphincterotomy did not remain patent. Doubilet illustrates how sphincteric activity can be maintained after sphincterotomy by residual action of the duodenal wall, acting as a constricting mechanism.[8] (See Fig. 12.) It then becomes conceivable how restenosis may occur. Sphincteroplasty, as advocated by Jones,[12] remains permanently patent. Demonstration of its patency is done by the reflux of barium into the biliary tree on upper GI. Lack of patency was demonstrated in one patient with a prior sphincterotomy, case #2, by the inability to demonstrate reflux on upper GI.

Typically, the patient with primary stenosis of the sphincter of Oddi will present with pain in the second or third decade of life. Oftentimes the patient will be suspected of having psychogenic pain, or has been subjected to multiple surgical procedures to find a cause for the pain. The patient may also give a history of a morphine allergy, whereby attempted use of this analgesic will have a paradoxical effect and bring on the pain. A history of alcohol abuse, in general, will be absent. The pain will suggest either acute cholecystitis or acute pancreatitis, characterized as being right upper quadrant or epigastric in nature with radiation to the back. There may be associated nausea and vomiting; fever and chills may accompany the pain.

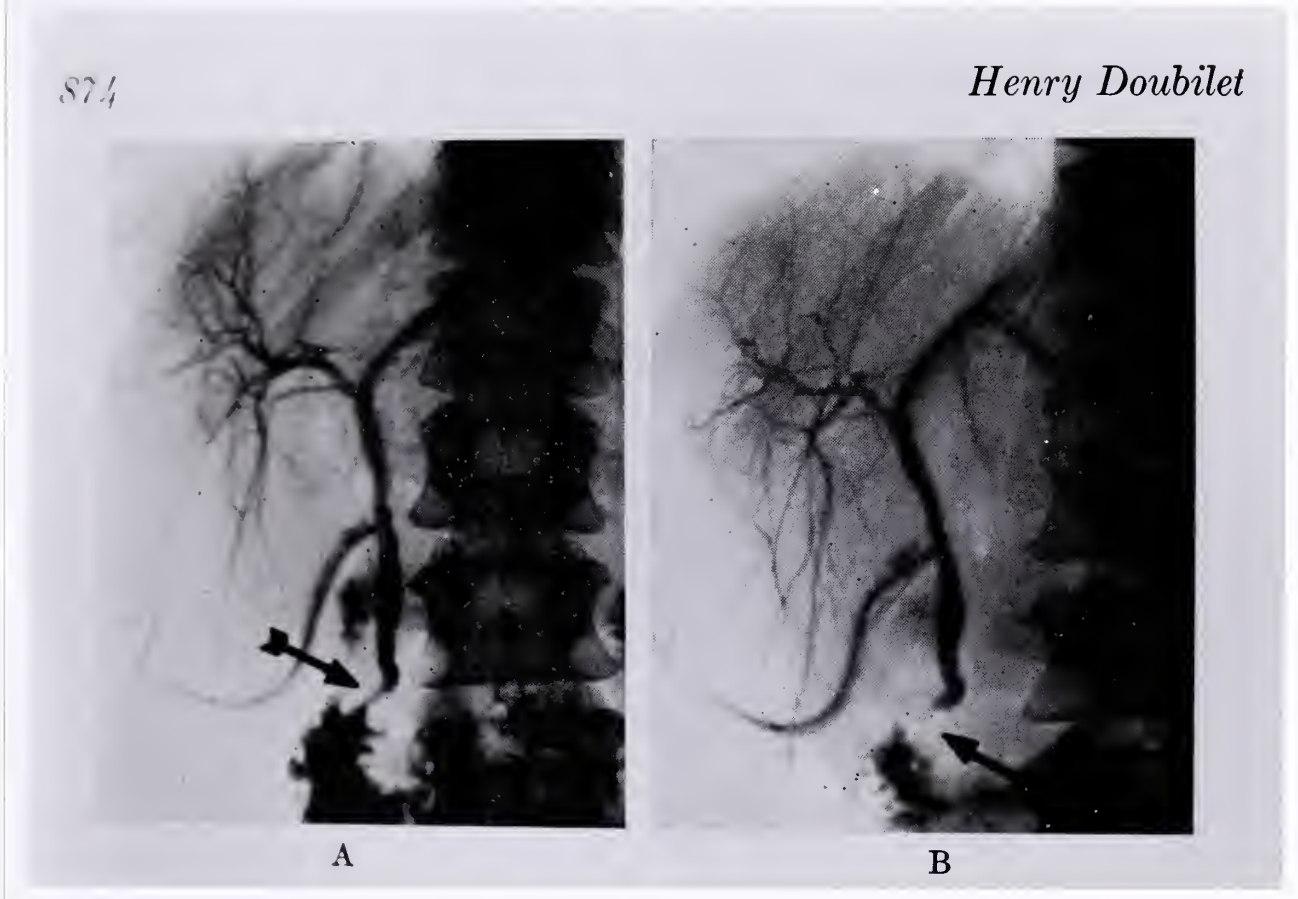


Fig. 12. Cholangiographic study done by Doubilet, demonstraring residual sphincteric activity after sphincterotomy. (A) Flow of dye into the duodenum through a sectioned sphincter, (B) five minutes after morphine administration, duodenal wall compresses the duct.[8]

Appropriate laboratory studies may show elevated amylase and lipase in the case of idiopathic pancreatitis. Cholestasis may not be reflected by abnormal liver function studies, jaundice not being a consistent finding. Procedures such as an oral cholecystogram, intravenous cholangiogram, or ERCP will aid in the diagnosis by exclusion, by demonstrating the absence of calculi in the common bile duct. They may also demonstrate pathology with the finding of grossly dilated pancreatic and common bile ducts. According to Cattell, et al.,[5] the criteria established by Wise and O'Brien for determining common bile duct obstruction on intravenous cholangiogram are: 1) a ductal diameter greater than 15 mm which invariably indicates obstruction, and 2) a diameter between 8-15 mm could conceivably be partial obstruction. Determination of partial obstruction is done using time - density relationships of the contrast material at 60 minutes and 120 minutes. The exception to this rule is in the case of a functioning gallbladder which prolongs the density.

The study presented made use of ERCP as a diagnostic tool. Cannulation of the papilla of Vater has been, until recently, an intraoperative procedure. ERCP proves to be a very useful tool in ascertaining, preoperatively, the pathology of the distal common bile duct. Stenosis may be suspected by observing retention of contrast material in the common bile duct. The extent

of pancreatic and common bile duct dilatation may also be determined. Currently at Yale, studies are being done to correlate manometric studies with variation of sphincter tone.[6] This may add more variables in the assessment of primary stenosis.

The laboratory methods just described aid the clinician by raising his index of suspicion for stenosis. The definitive procedure, of course, is exploratory laparotomy with direct visualization of the papilla of Vater. Stenosis is determined by subjecting it to the test of being able to accept the passage of a #3 Bakes dilator. Fibrotic thickening may be observed at this point , as may varying degrees of inflammation.

The criteria used in this study are intended to enable exclusion of other possible causes of stenosis of the sphincter of Oddi. Primary stenosis represents a seemingly de novo pathologic process. The importance of this lesion, until recently, lay in its ability to render medical science impotent to achieve resolution of the pain experienced by its patients. Clearly, sphincteroplasty will not benefit all patients with stenosis, especially those who may receive emotional gratification from the pain, but recognition of the possibility of the existence of this lesion will enable the clinician to fulfill his moral obligation to his patients.

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